Neighborhood Health Plan OF RHODE ISLAND™
910 Douglas Pike, Smithfield, RI 02917 : 1-800-963-1001 : nhpri.org

Spinraza® (nusinersen)
(Intrathecal)

Effective Date: 04/10/2019
Dates Reviewed: 04/10/2019, 7/26/2019, 1/15/2020, 8/24/2020, 01/12/2021, 01/20/2022, 2/23/2023, 12/07/2023, 01/04/2024, 03/12/2025, 3/10/2026

Scope: Medicaid, Commercial, Medicare

I. Length of Authorization

- Initial coverage criteria = 6 months
- Continuation of therapy = 12 months

II. Dosing Limits

- Initial dose: 120 billable units on day 0, day 14, day 28, day 59 (480 units)
- Renewal: 120 billable units every 120 days (360 units)

III. Summary of Evidence

Summary of Evidence: Spinraza is a survival motor neuron-2 (SMN2)-directed antisense oligonucleotide indicated for the treatment of spinal muscular atrophy (SMA) in pediatric and adult patients. Clinical trials evaluating the efficacy and safety of Spinraza in patients with SMA have demonstrated significant therapeutic benefits, including improved motor function, increased survival, and delayed disease progression. Common adverse events are mild to moderate injection site reactions and respiratory infections.

IV. Initial Approval Criteria

Submission of supporting clinical documentation (including but not limited to medical records, chart notes, lab results, and confirmatory diagnostics) related to the medical necessity criteria is REQUIRED on all requests for authorizations. Records will be reviewed at the time of submission as part of the evaluation of this request. Please provide documentation related to diagnosis, step therapy, and clinical markers (i.e., genetic, and mutational testing) supporting initiation when applicable. Please provide documentation via direct upload through the PA web portal or by fax. Failure to submit the medical records may result in the denial of the request due to inability to establish medical necessity in accordance with policy guidelines.

- Member must have the following laboratory tests at baseline and prior to each administration (laboratory tests should be obtained within several days prior to administration): platelet count, prothrombin time, activated partial thromboplastin time, and quantitative spot urine protein testing; AND
- Member retains voluntary motor function (e.g. manipulate objects using upper extremities, ambulate, etc.); AND
- Member must have a diagnosis of 5q spinal muscular atrophy confirmed by either homozygous deletion of the SMN1 gene or dysfunctional mutation of the SMN1 gene; AND
- Member has at least 2 copies of SMN2; AND
- Member has not received a dose of Zolgensma (onasemnogene abeparvovec-xioi) or Itvisma (onasemnogene abeparvovec-brve) in the past and will not be used concurrently with Spinraza (nusinersen); AND
- Member will not be using Spinraza (nusinersen) in combination with Evrysdi (risdiplam); AND
- Member is not dependent on either of the following:
    - Invasive ventilation or tracheostomy.
    - Use of non-invasive ventilation beyond the use for naps and nighttime sleep; AND
- Member must have a diagnosis of SMA phenotype I, II, or III; AND
    - Member has ≤ 3 copies of the *SMN2* gene; OR
    - Member has symptomatic disease (i.e., impaired motor function and/or delayed motor milestones); AND
- Baseline documentation of one or more of the following:
    - Motor function/milestones, including but not limited to, the following validated scales: Hammersmith Infant Neurologic Exam (HINE), Hammersmith Functional Motor Scale Expanded (HFMSE), Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP INTEND), 6-minute walk test (6MWT), upper limb module (ULM), etc.
    - Respiratory function tests (e.g., forced vital capacity [FVC], etc.).

- Exacerbations necessitating hospitalization and/or antibiotic therapy for respiratory infection in the preceding year/timeframe.
- Member weight (for members without a gastrostomy tube)

- Medicare members who have previously received this medication within the past 365 days are not subject to Step Therapy Requirements

V. Renewal Criteria

- Member meets all initial criteria; AND
- Absence of unacceptable toxicity which would preclude safe administration of the drug. Examples of unacceptable toxicity include: significant renal toxicity, thrombocytopenia, coagulation abnormalities, etc.; **AND**
- Member has not received a dose of Zolgensma (onasemnogene abeparvovec-xioi) or Itvisma (onasemnogene abeparvovec-brve) in the past and is not being used in combination with Evrysdi (risdiplam); AND
- Recent laboratory values (i.e. platelet count, prothrombin time, activated partial thromboplastin time, and quantitative spot urine protein testing) must be submitted associated with last dose given; AND
- Member has responded to therapy compared to pretreatment baseline (e.g., chart notes) by two or more of the following:

  - Prescriber must submit medical records (e.g., chart notes, laboratory values) with the most recent results documenting a positive clinical response from pretreatment baseline status to Spinraza therapy as demonstrated by at least one of the following exams:

    A. HINE-2 milestones:

    One of the following:

    - Improvement or maintenance of previous improvement of at least 2 point (or maximal score) increase in ability to kick.
    - Improvement or maintenance of previous improvement of at least 1 point increase in any other HINE-2 milestone (e.g., head control, rolling, sitting, crawling, etc.) excluding voluntary grasp; AND

    One of the following:

    - The member exhibited improvement or maintenance of previous improvement in more HINE motor milestones than worsening, from pretreatment baseline (net positive improvement).
    - Achieved and maintained any new motor milestones when they would otherwise be unexpected to do so (e.g., sit unassisted, stand, walk); OR

B. HFMSE:

One of the following:

- Improvement or maintenance of previous improvement of at least a 3 point increase in score from pretreatment baseline.
- Member has achieved and maintained any new motor milestone from pretreatment baseline when they would otherwise be unexpected to do so; OR

C. ULM:

One of the following:

- Improvement or maintenance of previous improvement of at least a 2 point increase in score from pretreatment baseline.
- Member has achieved and maintained any new motor milestone from pretreatment baseline when they would otherwise be unexpected to do so; OR

D. CHOP INTEND:

One of the following:

- Improvement or maintenance of previous improvement of at least a 4 point increase in score from pretreatment baseline.
- Member has achieved and maintained any new motor milestone from pretreatment baseline when they would otherwise be unexpected to do so;

- Stability or improvement in respiratory function tests (such as forced vital capacity [FVC], etc.)
- Reductions in exacerbations necessitating hospitalization and/or antibiotic therapy for respiratory infection in the preceding year/timeframe
- Stable or increased weight (for member's without a gastrostomy tube)
- Slowed rate of decline in the aforementioned measures

The following HCPCS/CPT codes are:

HCPCS/CPT Code	Description
J2326	Injection, nusinersen, 0.1mg

References:

1. Spinraza prescribing information. Cambridge, MA.: Biogen, Inc.; April 2024. accessed December 2025.
2. Markowitz JA, Singh P, Darras BT. Spinal Muscular Atrophy: A Clinical and Research Update. Pediatric Neurology 46 (2012) 1-12.

3. Sugarman EA, Nagan N, Zhu H, et al. Pan-ethnic carrier screening and prenatal diagnosis for spinal muscular atrophy: clinical laboratory analysis of >72,400 specimens. Eur J Hum Genet 2012;20:27-32.
4. Prior TW, Snyder PJ, Rink BD, et al. Newborn and carrier screening for spinal muscular atrophy. Am J Med Genet A. 2010 Jul;152A(7):1608-16.

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
G12.0	Infantile spinal muscular atrophy, type I [Werdnig-Hoffmann]
G12.1	Other inherited spinal muscular atrophy
G12.25	Progressive spinal muscle atrophy
G12.8	Other spinal muscular atrophies and related syndromes
G12.9	Spinal muscular atrophy, unspecified

Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

The preceding information is intended for non-Medicare coverage determinations. Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determinations (NCDs) and/or Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. Local Coverage Articles (LCAs) may also exist for claims payment purposes or to clarify benefit eligibility under Part B for drugs which may be self-administered. The following link may be used to search for NCD, LCD, or LCA documents: https://www.cms.gov/medicare-coverage-database/search.aspx. Additional indications, including any preceding information, may be applied at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes		
Jurisdiction	**NCD/LCA/LCD Document (s)**	**Contractor**
E, F	A58578	Noridian Healthcare Solutions, LLC

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
E (1)	CA, HI, NV, AS, GU, CNMI	Noridian Healthcare Solutions, LLC
F (2 & 3)	AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ	Noridian Healthcare Solutions, LLC
5	KS, NE, IA, MO	Wisconsin Physicians Service Insurance Corp (WPS)
6	MN, WI, IL	National Government Services, Inc. (NGS)
H (4 & 7)	LA, AR, MS, TX, OK, CO, NM	Novitas Solutions, Inc.
8	MI, IN	Wisconsin Physicians Service Insurance Corp (WPS)
N (9)	FL, PR, VI	First Coast Service Options, Inc.
J (10)	TN, GA, AL	Palmetto GBA
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA)	Novitas Solutions, Inc.
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)
15	KY, OH	CGS Administrators, LLC

Policy Rationale:

Spinraza was reviewed by the Neighborhood Health Plan of Rhode Island Pharmacy & Therapeutics (P&T) Committee. Neighborhood adopted the following clinical coverage criteria to ensure that its members use Spinraza according to Food and Drug Administration (FDA) approved labeling and/or relevant clinical literature. Neighborhood worked with network prescribers and pharmacists to draft these criteria. These criteria will help ensure its members are using this drug for a medically accepted indication, while minimizing the risk for adverse effects and ensuring more cost-effective options are used first, if applicable and appropriate. For Medicare members, these coverage criteria will only apply in the absence of National Coverage Determination (NCD) or Local Coverage Determination (LCD) criteria. Neighborhood will give individual consideration to each request it reviews based on the information submitted by the prescriber and other information available to the plan.